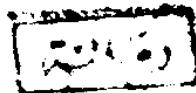


CONGENITAL ANOMALIES OF THE URINARY BLADDER

Essay

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Master Degree of (*Urology*)

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INTRODUCTION

Many of the congenital anomalies of the urinary bladder are rare and a few cases have been described in the literature. In order to understand the complex embryogenesis of vesical anomalies, the development of the urinary bladder and the urethra are discussed as an introduction to the congenital anomalies of the urinary bladder.

Besides curiosity and the desire for knowledge for its own sake, there are important other reasons to study the congenital bladder anomalies. The greatest practical reason is the hope of improving the miserable life of the patients with these deformities. Untreated patients certainly lead wretched, piteous lives. Improvements in the methods of management have undoubtedly lessen the burdens of these cripples.

DEVELOPMENT OF THE URINARY BLADDER & URETHRA

EMBRYOLOGY OF THE URINARY BLADDER

The urinary bladder is derived partly from the endodermal cloaca and partly from the caudal ends of the mesonephric ducts (*Warwick and Williams, 1973*).

The Cloaca: (Figs. 1, 2)

The cloaca (Latin - a sewer) is appropriately named as it is a cavity common to both urogenital and alimentary system into which open, in the early stages of development, the allantois and the hindgut. Its lumen is separated from the amniotic cavity by a thin cloacal membrane, which is present much earlier in development as a small area into which mesoderm for the primitive streak does not penetrate so that the endoderm and ectoderm come into contact.

The allantois is a blind-ended diverticulum of endoderm that projects into the mesoderm of the body stalk (connecting stalk). It is vestigial and associated with the development of the excretory system.

The dorsal part of the cloaca forms the lower end of the hindgut and the ventral part, which merges imperceptibly into the allantois, becomes the primitive urogenital sinus. As the "spur" between the cavities deepens and grows caudally, it approaches the cloacal membrane and will eventually completely separate the hindgut from the primitive urogenital sinus. It is therefore known as the urorectal septum. This occurs at about 6 weeks of intra-uterine life (*Moffat, 1984*).

At about the eighth week the cloacal membrane ruptures, separation of the cloaca is complete and its two parts open independently. The opening

of the rectum is the anus, and that of the urogenital sinus is the ostium urogenitale.

Meanwhile, the proximal part of the allantois has become greatly dilated and may now quite properly be called the urinary bladder. The neck of the bladder has been formed from tissue which was originally part of the cloaca.

The apex of the vesico-urethral canal is prolonged to the umbilicus as a narrow canal termed the **urachus**. In post-natal life, the urachus is drawn downwards as the bladder descends, but its blind upper end remains connected to one or both of the obliterated umbilical arteries. Its lumen persists throughout life, and its lower end frequently communicates with the bladder near its apex (*Warwick and Williams, 1973*).

The urachus is contiguous with the allantoic duct, but the allantois probably does not contribute to the formation of the urachus or bladder (*Felix, 1912; Arey, 1974*).

By 12 weeks, the segment of the bladder contiguous with the allantoic diverticulum involutes. This segment is between the apex of the bladder and the umbilicus. It becomes a thick tube and then a fibrous cord, the median umbilical ligament (*Moore, 1977*).

The Mesonephric Ducts

The mesonephric ducts open from either sides into the cephalic part of the cloaca. After the urorectal fold has divided the cloaca, the mesonephric ducts appear to empty into the allantois. The mesonephric ducts are actually openings into the urogenital sinus which is continuous with the allantois.

In the growth of the bladder, the caudal portion of the mesonephric duct is absorbed into the bladder wall. This absorption progress until the part of the mesonephric duct caudal to the point of origin of the metanephric diverticulum has disappeared. The end result of this process is that the mesonephric and metanephric ducts open independently into the urogenital sinus. The mesonephric ducts open into the part of the urogenital sinus which remains narrower and gives rise to the urethra (*Bradley, M., and Bruce, M., 1974*).

By 28 days of development, the mesonephric ducts have already reached and fused with the urogenital sinus. This fusion involves the coalescence of the epithelium of the urogenital sinus (endodermal derivative) with that of the mesonephric duct (mesoderm derivative). At this time the ureter originates from mesonephric duct. The segment of the mesonephric duct distal to the site of origin of the ureteral bud dilates as the common excretory duct is absorbed into the urogenital sinus either directly or after terminus of the duct loop (*Hamilton and Mossman, 1976b*).

After the right and the left common excretory ducts have been absorbed into the urogenital sinus, the epithelia of the ducts fuse towards the midline as a triangular area, **the primitive trigone** (*Max Maizels, 1986*).

In this way, the mesonephric duct contributes to the trigone of the bladder and dorsal wall of the prostatic urethra. The remainder of the vesico-urethral part forms the body of the prostatic urethra (*Warwick and Williams, 1973*).

The mesonephric ducts also migrate caudally and flank the paramesonephric ducts at the level of the urogenital sinus. This is the site of the future verumontanum (*Max Maizels, 1986*).

The Urogenital Sinus

The urogenital sinus can be divided into 2 main segments; the dividing line is the junction of the combined müllerian ducts with the urogenital sinus (Müller's tubercle). The segments are as follows:

1. The ventral and pelvic portion will form the bladder, part of urethra in the male, and the whole urethra in the female. This portion receives the ureter.
2. The urethral or phallic portion receives the mesonephric and fused müllerian ducts. This will be part of urethra in the male and forms the lower fifth of the vagina and vaginal vestibule in the female (*Smith.D., 1981*).

The urorectal septum grows down between the primitive urogenital sinus and the hindgut until it reaches the cloacal membrane. The septum thus separates the gut from the developing urogenital system and divides the cloacal membrane into an anal and a urogenital membrane (Fig. 1).

The urogenital membrane extends as far forwards and cranially as attachment of the umbilical cord so that at this stage it faces forwards and there is no infra-umbilical abdominal wall.

The primitive urogenital sinus now changes its shape somewhat, becoming subdivided into a rather cylindrical vesico-urethral canal above the level of the openings of the mesonephric ducts and a definitive urogenital sinus below these openings.

The definitive urogenital sinus is subdivided into a short narrow cylindrical portion, the pelvic part (*pars pelvina*) and a more extensive phallic part (*pars phallica*) which is flattened from side to side (Fig. 3).

Mesoderm from the most caudal part of the primitive streak grows forwards on either side of the urogenital membrane and proliferates to form a pair of genital swellings, while anteriorly this mesoderm passes between the endoderm and the ectoderm of the cloacal membrane to form a pair of swellings covered with ectoderm, which soon fuse to form a single mid-line eminence known as the genital tubercle (Fig. 4). Behind this is therefore placed the urogenital membrane between the two genital swellings while in front and above it is the attachment of the umbilical cord, for there is still no infra-umbilical anterior abdominal wall.

A little later, however, this part of the abdominal wall develops by the ingrowth of more mesodermal cells between the endoderm and ectoderm and much later developing muscle cells from the mesodermal somites will invade this mesoderm to form the abdominal musculature.

It is important to realize that if mesoderm does not grow forwards from the primitive streak to form the genital tubercle and the infra-umbilical part of the abdominal wall there will be a region below the umbilicus where a membrane, the original cloacal membrane, composed only of ectoderm and endoderm, will exist.

It is the formation of the infra-umbilical part of the abdominal wall that rotates the plane of the cloacal membrane so that it faces, downwards, between the legs rather than forwards below the umbilicus.

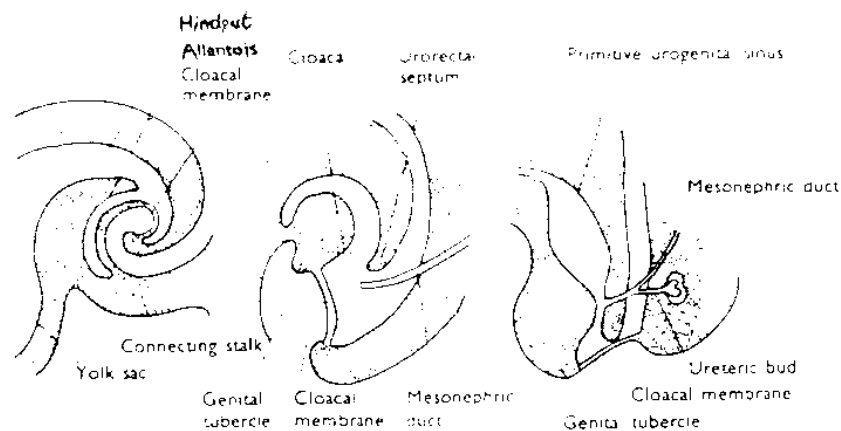


Fig. 1. The splitting of the cloaca by the urorectal septum to form the primitive urogenital sinus.

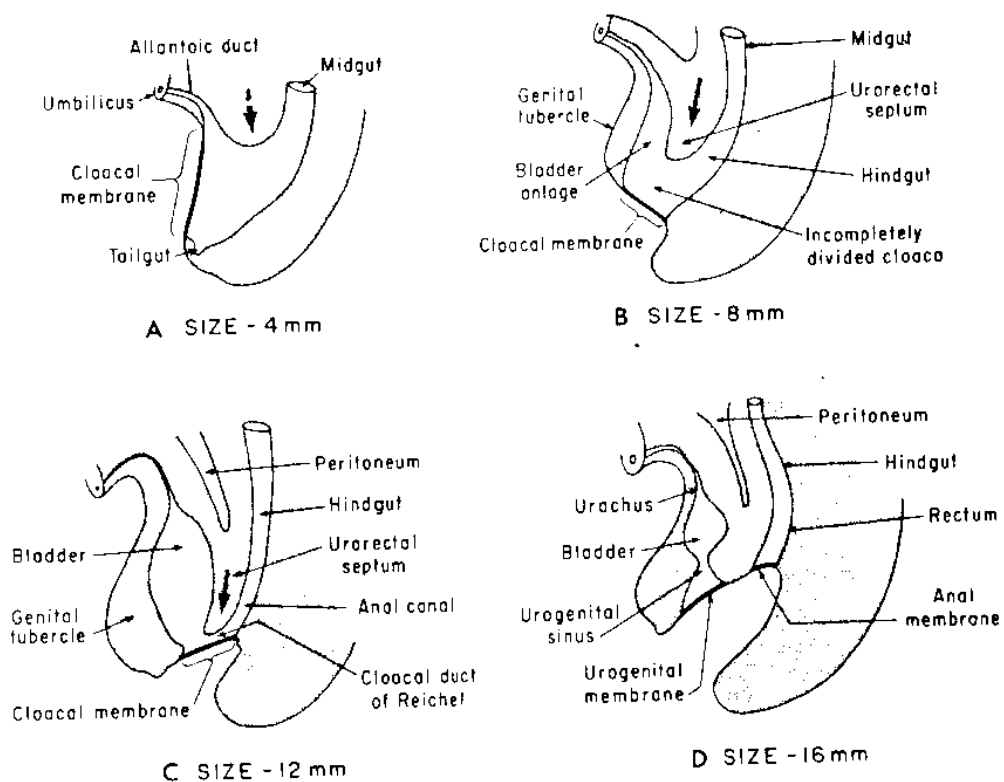


Fig. 2. Developmental changes of the cloaca and cloacal membrane in the 4-mm to 16-mm embryo. Arrows show direction of growth of the urorectal septum.

The genital tubercle is large and is recognizable as a phallus. Medial to the genital swellings is a pair of thin urethral folds that flank the opening of the urogenital sinus. The urogenital membrane have disintegrated.

The phallic part of the urogenital sinus extends forwards onto the ventral surface of the genital tubercle and is continuous further forwards with an ectodermal groove. This is called the urethral groove and it is limited on either side by a forward prolongation of the urethral folds.

The Bladder (Fig. 3)

The bladder and the upper part of the male urethra (the whole urethra in the female) develops from the endodermal vesico-urethral canal and from the lower end of the mesonephric duct including the origin of the ureteric bud which is, of course, mesodermal.

The manner in which the lower end of the mesonephric duct is taken into the vesico-urethral canal is not clearly understood but when this process has been completed the two mesonephric ducts open into the lower end of the vesico-urethral canal while the two ureters open into the wider upper part.

The vesico-urethral canal is by now clearly subdivided into a dilated segment which will form the bladder and a much narrower urethral portion, the ureters opening into the bladder at each upper corner of a well demarcated trigone.

A very plausible explanation of the process is that after the mesonephric duct and the ureter have been taken into the bladder, the lower end of the duct forms a caudally directed loop which is closely applied to the wall of the vesico-urethral canal.

The ascending limb of this loop then becomes absorbed into the wall of the vesico-urethral canal leaving the opening of the mesonephric duct at a much lower level (*Frazer, 1940 and Gyllensten, 1949*).

Other authors have described a dilatation at the lower end of the mesonephric duct which involves the orifice of the ureteric bud and which is absorbed into the bladder to form the trigone.

Whatever the precise mechanism of the absorption of the mesonephric duct into the urogenital sinus, the result of the process is that the opening of the ureteric bud into the mesonephric duct moves into the lower end of the vesico-urethral canal and then migrates upwards and laterally until it reaches its definitive position at the upper and outer angle of the trigone. The mesonephric duct itself remains as an opening at the lower end of the vesico-urethral canal (Fig. 3).

After the ureteric orifice has reached its definitive position it remains closed by a double-layered membrane, the lower layer of which is composed of epithelial cells similar to those of the bladder while the upper layer is continuous with the ureteric epithelium. This membrane is usually known as Chwalla's membrane although *Chwalla (1927)* himself called it the "Ureteren-membrane". It ruptures sometimes between the 25 and 30 mm. stage.

The presence of the membrane was confirmed by *Gyllensten (1949)* who found that its rupture was preceded by a dilatation of the lower end of the ureter, perhaps as a result of the secretion of urine by the metanephros.

The trigone itself, and the posterior wall of the urethra down as far as the openings of the mesonephric ducts (later the openings of the ejaculatory

ducts) are thus of mesodermal origin while the rest of the bladder is endodermal (Fig. 3).

This difference in developmental origin probably explains the structural differences between the trigone and the rest of the bladder. The trigonal musculature develops separately from that of the rest of the bladder and is derived from the ureteral musculature itself and from a deep periureteral sheath that surrounds the ureter (*Itatani et al., 1977*).

The apex of the urogenital sinus is continuous with the allantois and no distinct dividing line can be seen.

The lumen of the allantois becomes obliterated in the umbilical region at an early stage.

The vesico-urethral canal, however, extends up to this region to form the primitive bladder.

Obliteration continues in the cranial part of this so that the bladder apex gradually descends into the lower abdomen, leaving a fibrous band, the urachus, extending from the apex of the bladder to the umbilicus. This retains parts of its lumen into adult life. It is lined by transitional epithelium and may have cystic dilatations along its course. Its lower end often communicates with the lumen of the bladder (*Hinman, 1961*).

The bladder does not descend fully to its adult position until puberty. At birth the bladder neck is at the level of the upper border of the pubic symphysis so that the bladder is an abdominal rather than a pelvic organ (*Moffat, 1984*).